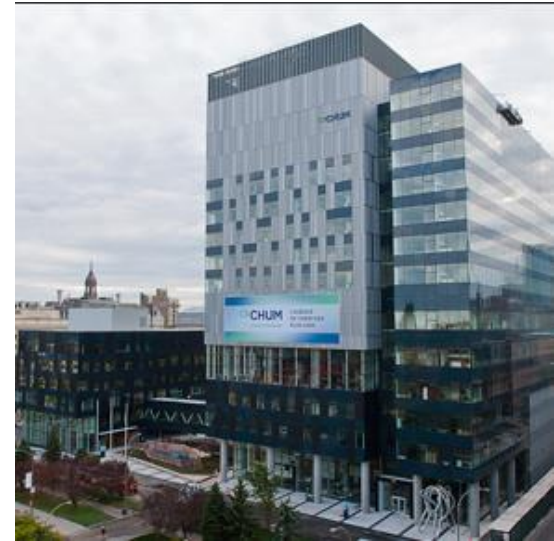


# Vascular malformation (VM) classification, clinical and typical imaging features

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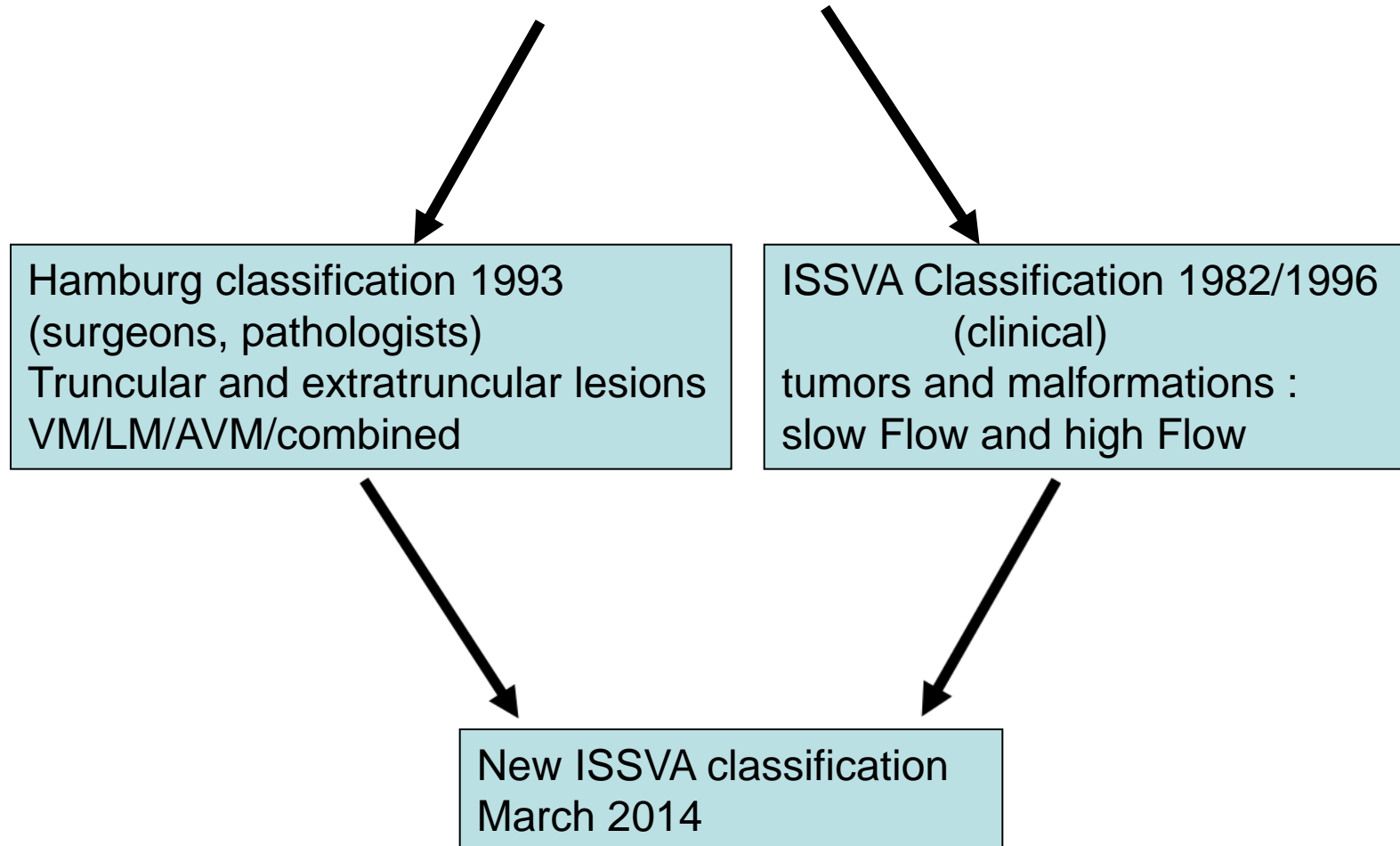
# Disclosure

- Research grant
  - Siemens Medical
  - Object Research System
  - Bracco Diagnostic
  - Biotronik
  - TVA medical
- Consultant
  - Cook Medical

# Basic principles

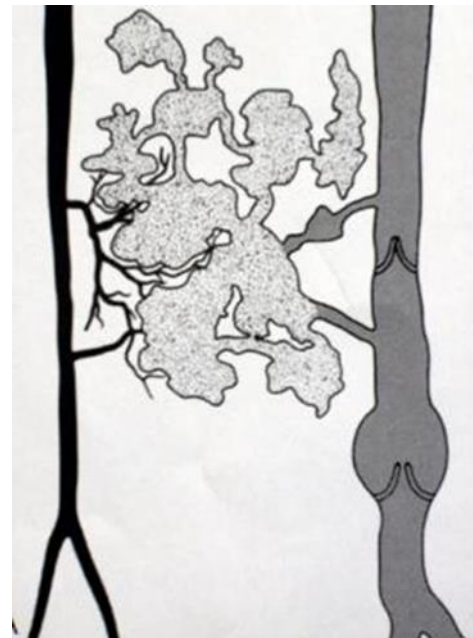
- Use an appropriate terminology
  - Mulliken classification + new ISSVA classification
- Always correlate imaging findings with clinical history and examination
  - Vascular anomalies clinic
- Multidisciplinary management
  - IR (diagnosis, intervention)
  - Plastic surgery, dermatology, ENT, internal medicine

# CLASSIFICATIONS



# Extratruncal vs truncal

- Extratruncal
  - Defect occurring early in the embryogenesis
  - Mesenchymal cell memory (proliferation)
  - No or few connections with normal vascular system
- Truncal
  - Defect occurring later
  - Hypoplasia or dilatation of the vascular system

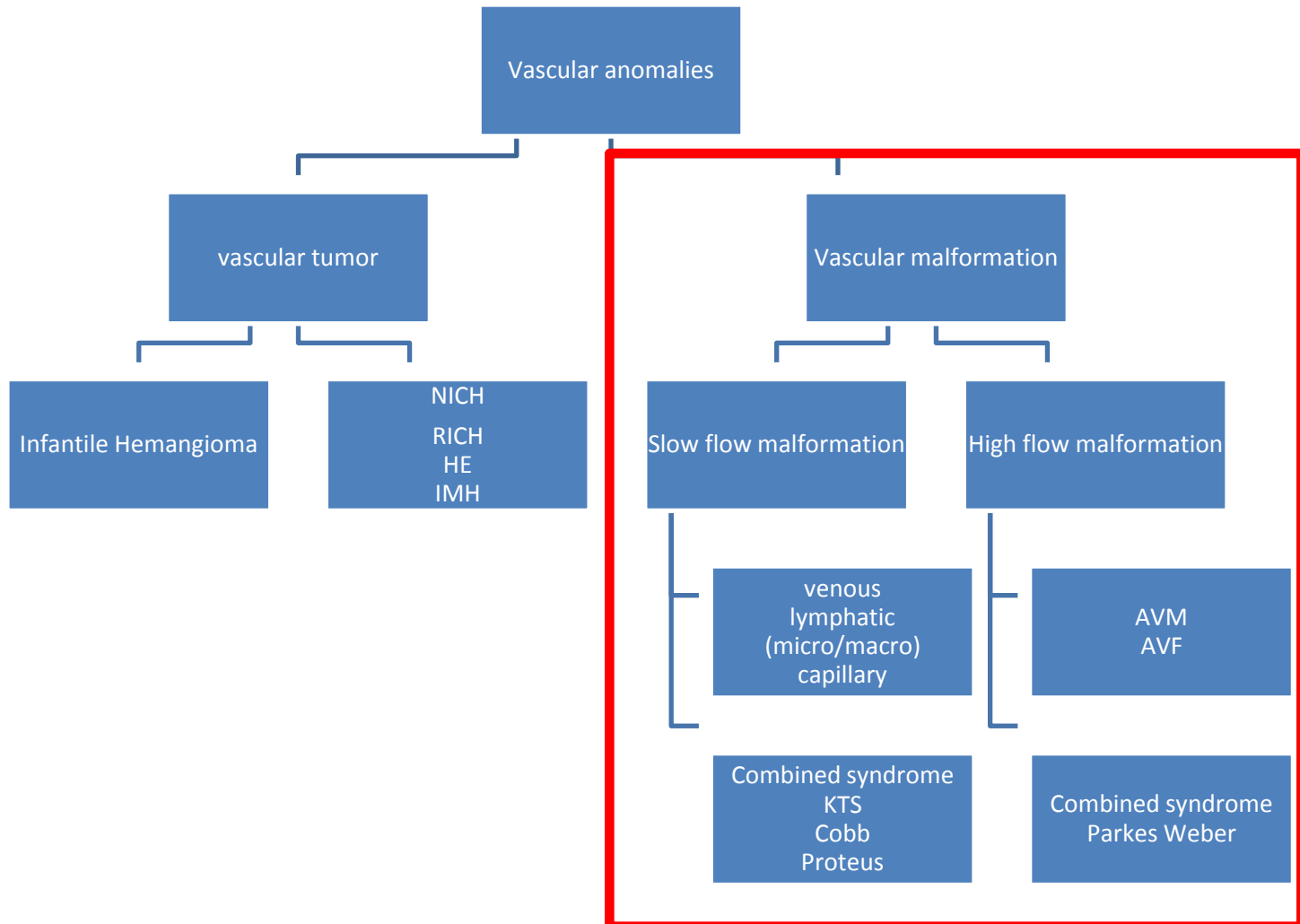


Extra truncal



Truncal

# Mulliken classification



Mulliken J. and Glowacki J.: Hemangiomas and vascular malformations in infants and children : a classification based on endothelial characteristics. *Plast Reconstr Surg* 1982; 69 : 412-422

# 2014 ISSVA classification

| Vascular anomalies  |  |  |                         |                                 |
|---|--|--|-------------------------|---------------------------------|
| Vascular tumors   | Vascular malformations   |  |                         |                                 |
|   | Simple   | Combined °   | of major named vessels  | associated with other anomalies |
| <a href="#">Benign</a><br><a href="#">Locally aggressive or borderline</a><br><a href="#">Malignant</a> | <a href="#">Capillary malformations</a><br><a href="#">Lymphatic malformations</a><br><a href="#">Venous malformations</a><br><a href="#">Arteriovenous malformations*</a><br><a href="#">Arteriovenous fistula*</a> | <a href="#">CVM, CLM</a><br><a href="#">LVM, CLVM</a><br><a href="#">CAVM*</a><br><a href="#">CLAVM*</a><br><a href="#">others</a> | <a href="#">Details</a> | <a href="#">Details</a>         |

# **Anomalies of major named vessels**

(aka "channel type" or "truncal" vascular malformations)

## Affect

lymphatics

veins

arteries

## Anomalies of

origin

course

number

length

diameter (aplasia, hypoplasia, stenosis, ectasia / aneurysm)

valves

communication (AVF)

persistence (of embryonal vessel)

# Investigation

- Clinical examination combined with DUS
  - Coloration
  - Softness
  - Thrill, expansion with valsalva
  - Vessel dilatation, venous reflux
  - Limb overgrowth
  - CM (angioma)
  - Tissular destruction

# Venous malformation

- Low flow
- Most frequent
  - Head and neck 40%
  - Body 20%
  - Limbs 40%
- Expansion
  - Valsalva
  - Dependent position
- Bluish coloration



# Investigation

- Coagulation work-up for large VM
  - 88% have a localized intravascular coagulopathy (LIC)<sup>1</sup>
- Doppler ultrasound
- MRI if a treatment is considered or diagnostic unclear
- TDM not contributive
- Angiography not necessary +++

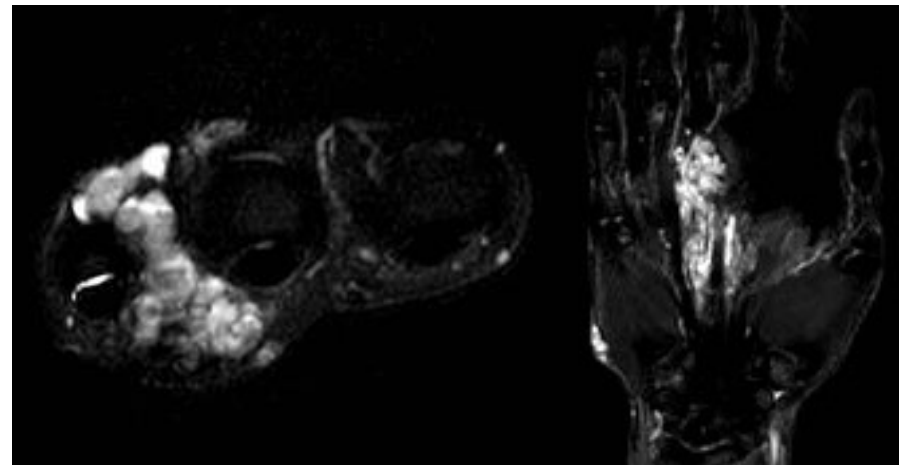
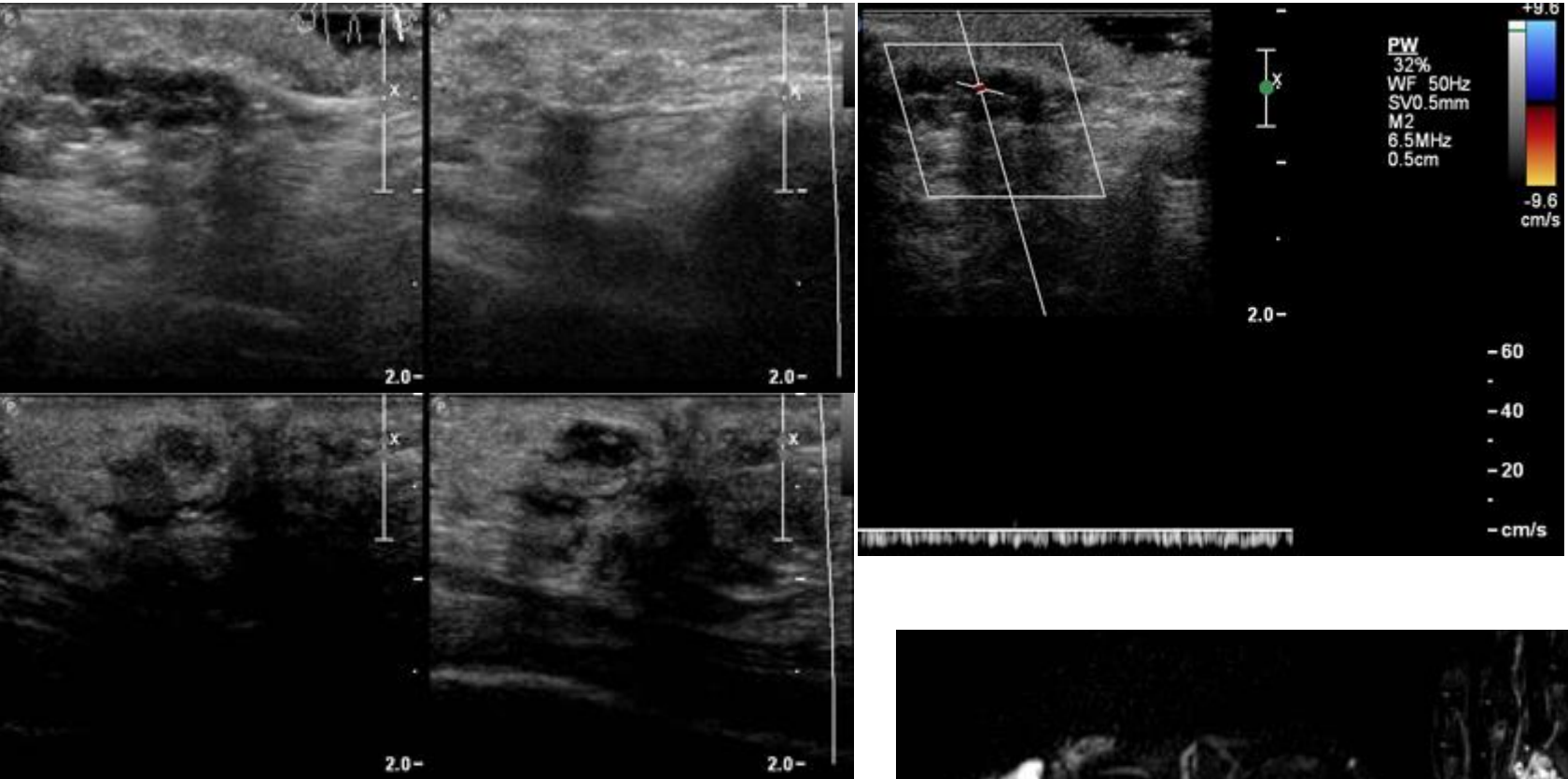
1. Enjolras O, J Am Acad Dermatol 1997;36:219–25.

# Venous malformation

- Doppler ultrasound
  - Low flow
  - Hypoechoic
  - Compressible venous dilatation
  - Phlebolitis
  - Evaluate feasibility of needle guidance for sclerotherapy

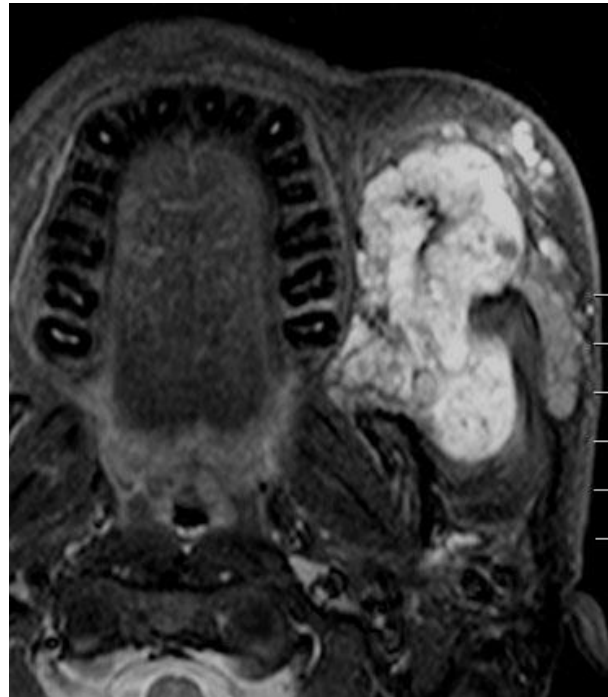


# Compressibility B-mode & Doppler

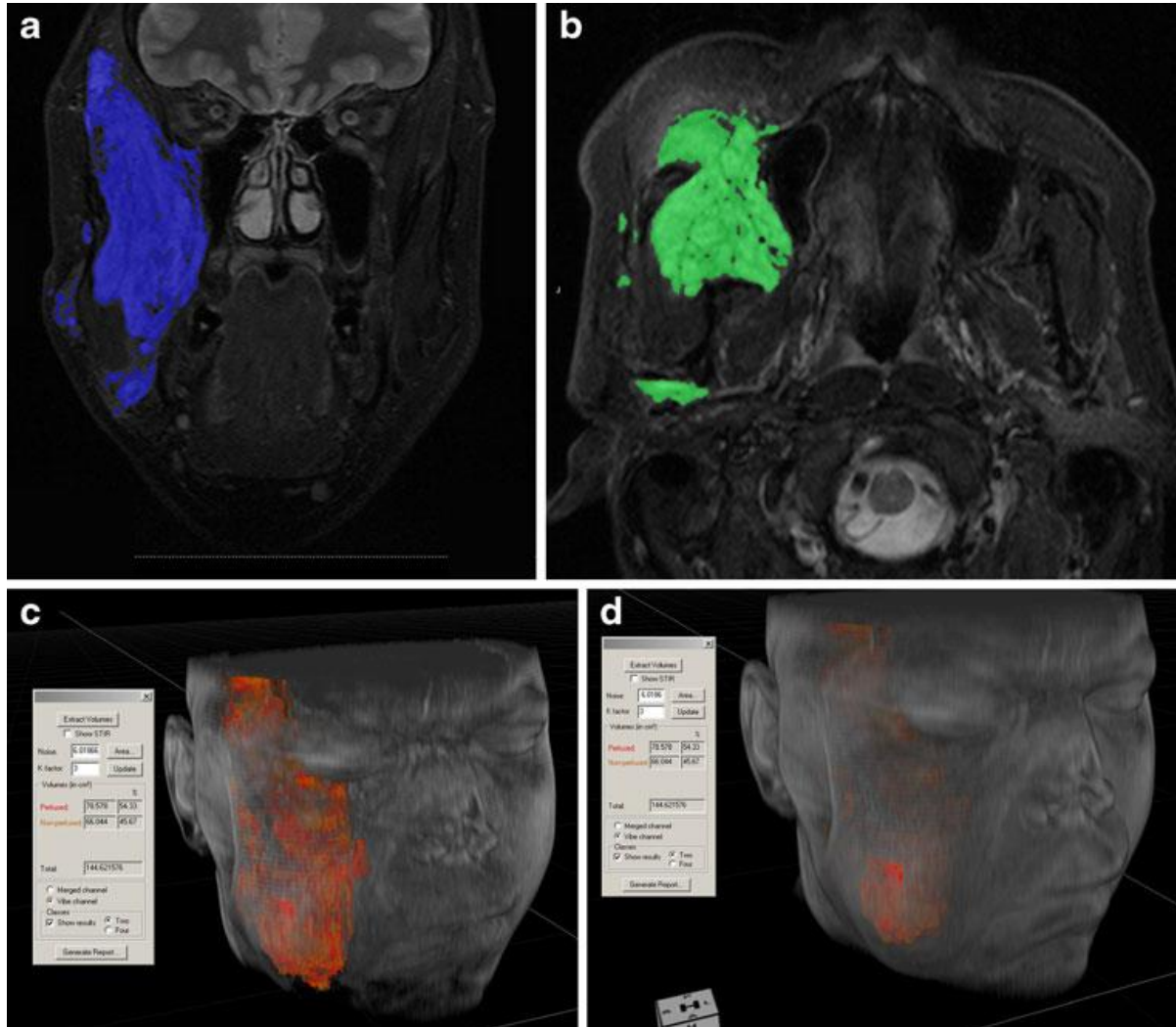


# VM & MRI

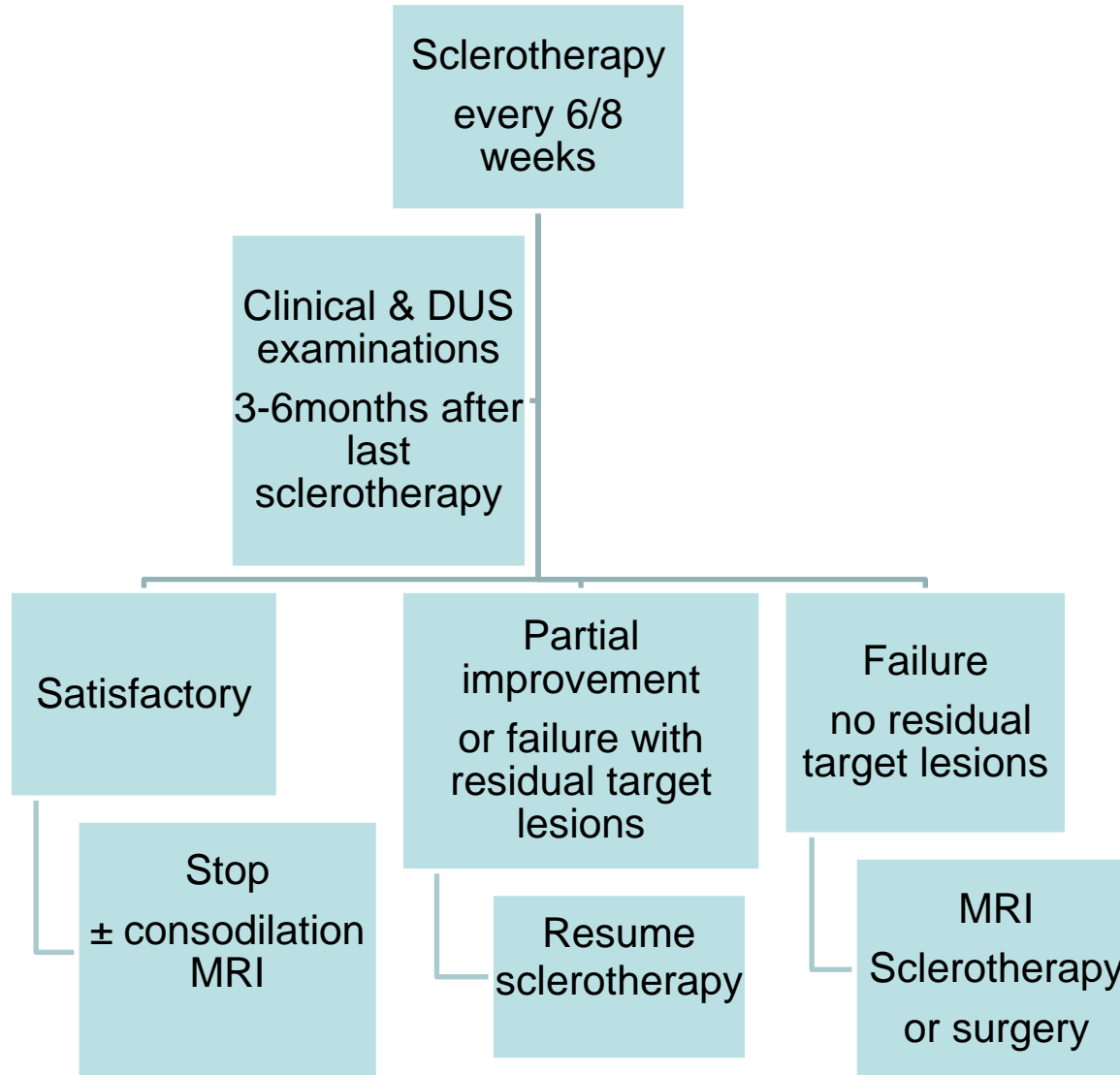
- Best examination for extension
- T2 (STIR), T1 and T1 fat sat post gado



# 3D modeling

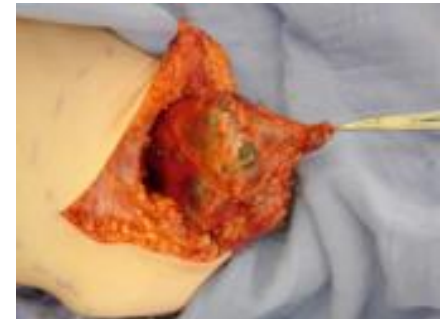


# VM follow-up



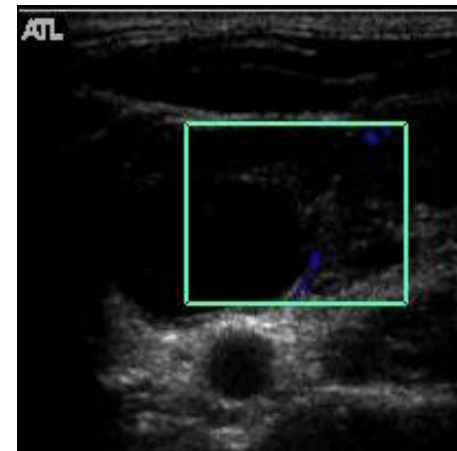
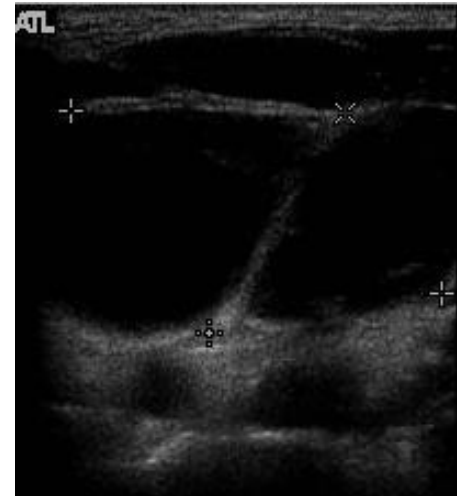
# Lymphatic malformation

- Cystic cavity lined by an endothelial layer filled by a lymphatic fluid
  - ML macrocystic ( $> 2\text{cm}^3$ )
  - ML microcystic ( $< 2\text{cm}^3$ )
  - Mixed lesion (micro-macro)
  - Mixed lesion lymphatic and venous
- Present at birth
- Growth childhood-teenage

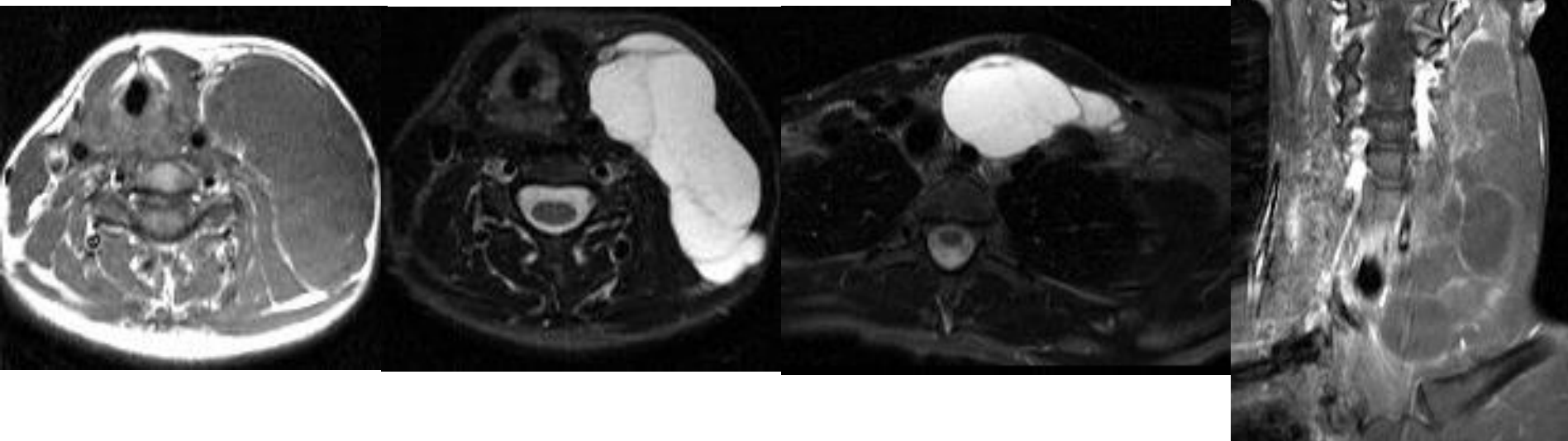


# Lymphatic malformation

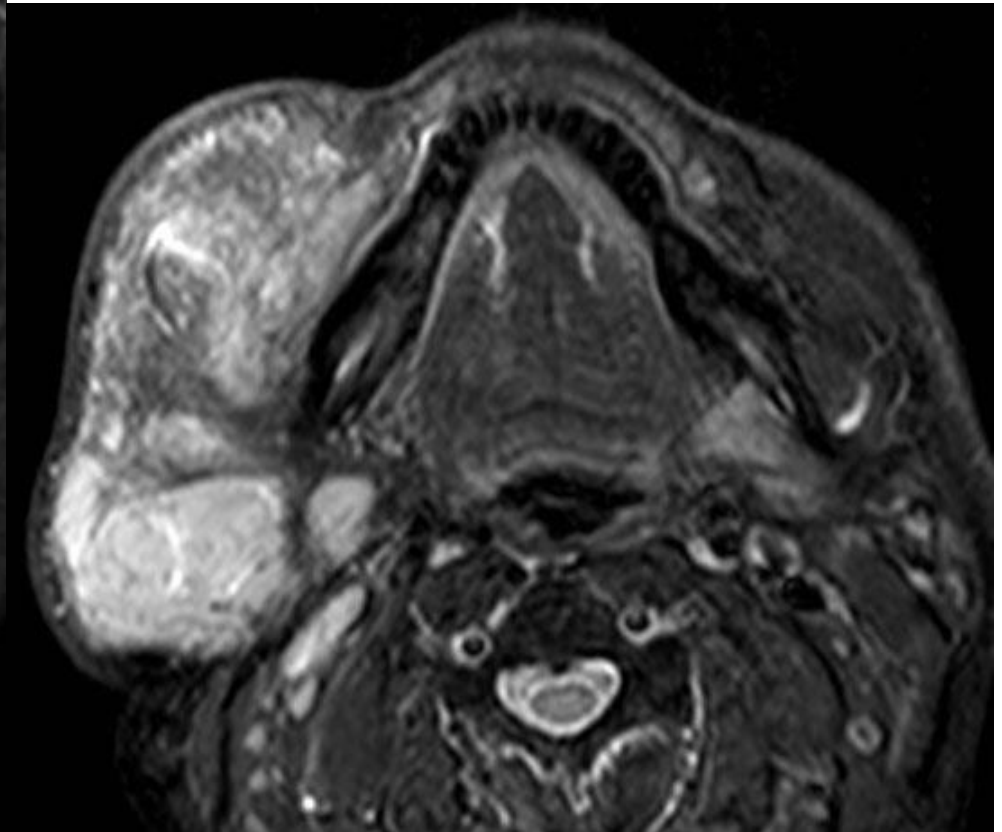
- Doppler ultrasound
  - Macrocyst with septation
  - Doppler
    - No flow in the cyst
    - High resistance flow in septa
  - Microcystic lesions infiltration of soft tissue by hyperechoic or heterogeneous mass



# MRI & LM

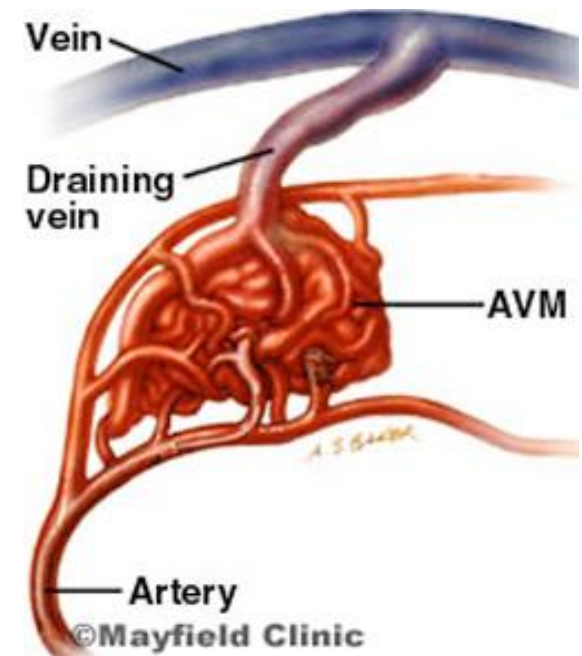


# Microcystic lymphangioma



# Arterio-venous malformation

- High-flow malformation
  - AV-shunting
  - Nidus
- Congenital
  - Expansion
    - Teenage
    - Pregnancy

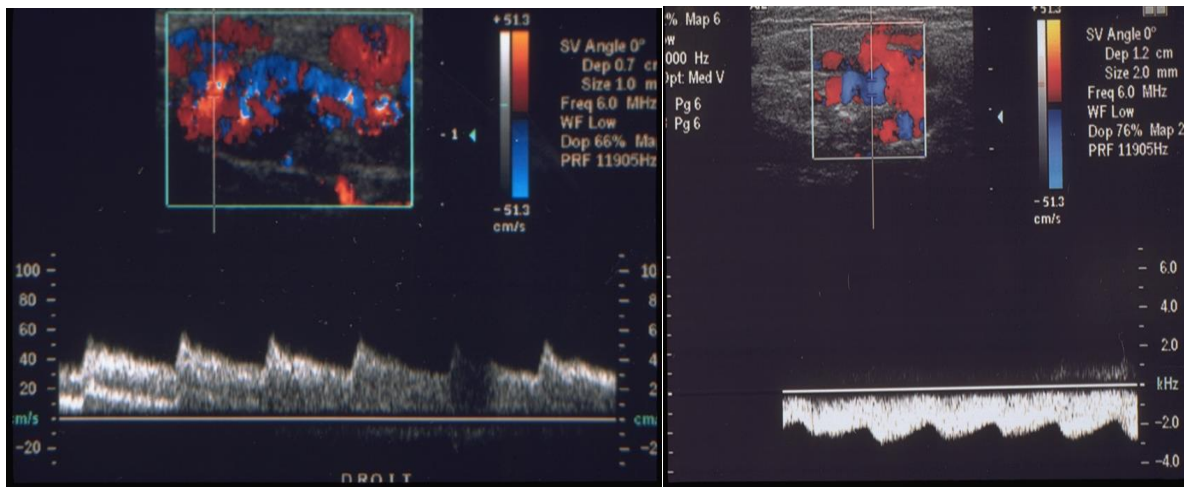


# Schobinger classification

(Clinical staging system to grade the evolution of AVMs)

## Stage 1: Quiescent

- No evidence of progression
- No treatment indication



# Schobinger classification

## Stage 2: Expansion

### Stage 1

+

- Darkening blush stain
- ↑ Pulsations
- ↑ Thrill
- ↑ Bruit
- ↑ Venous dilatation



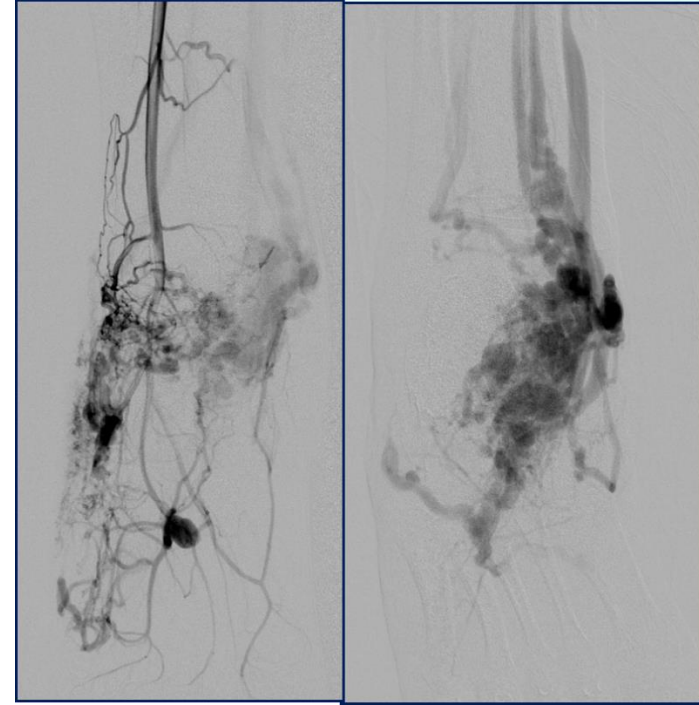
# Schobinger classification

## Stage 3: Destruction

### Stage 2

+

- Steal
- Distal ischemia
- Dystrophic skin changes
- Ulceration
- Bleeding
- Persistent pain
- Tissues necrosis
- Soft tissues and bones changes

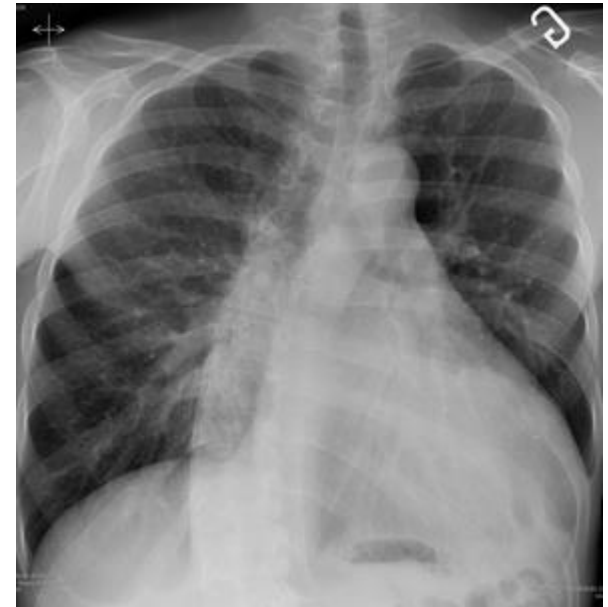


# Schobinger classification

## Stage 4: Decompensation

**Stage 3 +**

- High output cardiac failure



# Doppler +++

## High flow in vascular nidus

### – Arterial side

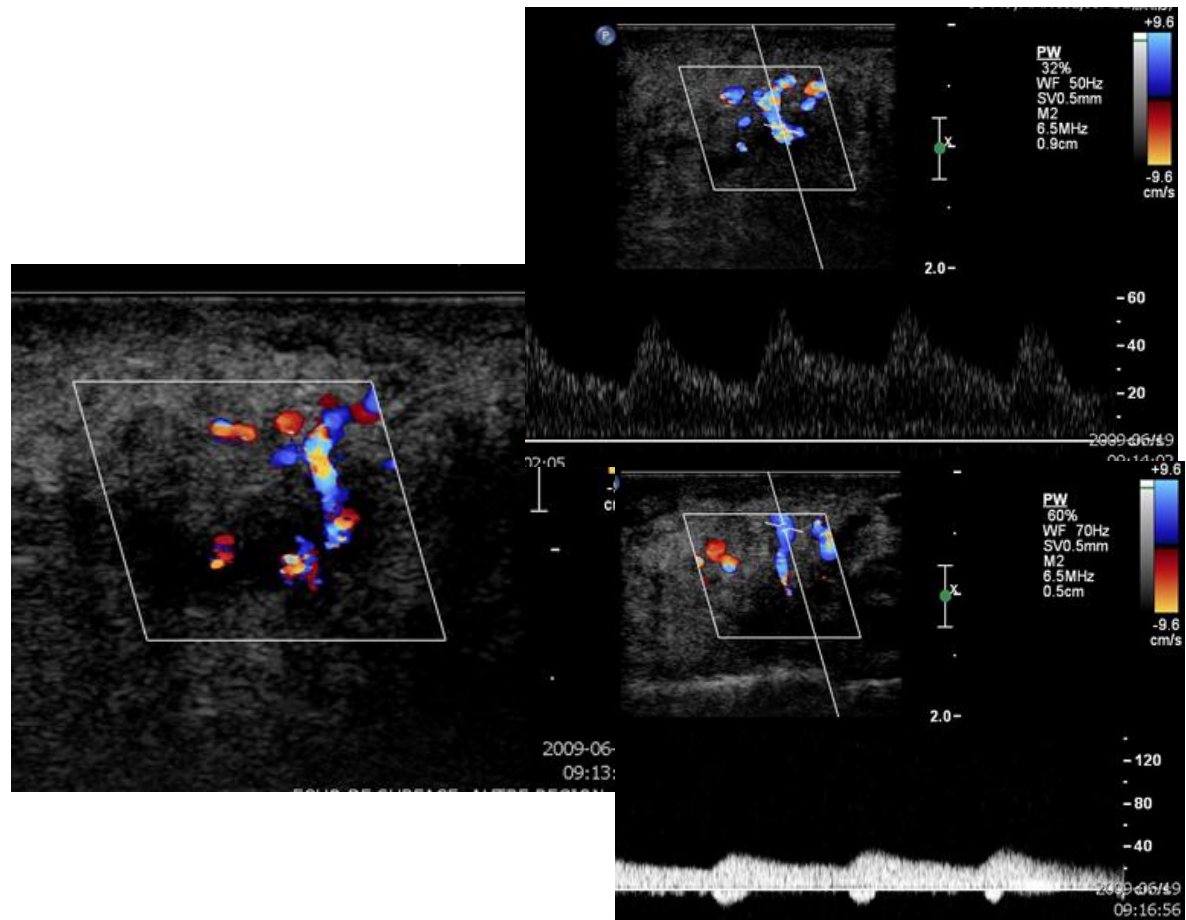
- High velocities and diastolic flow

### – Venous side

- Arterialization of venous flow

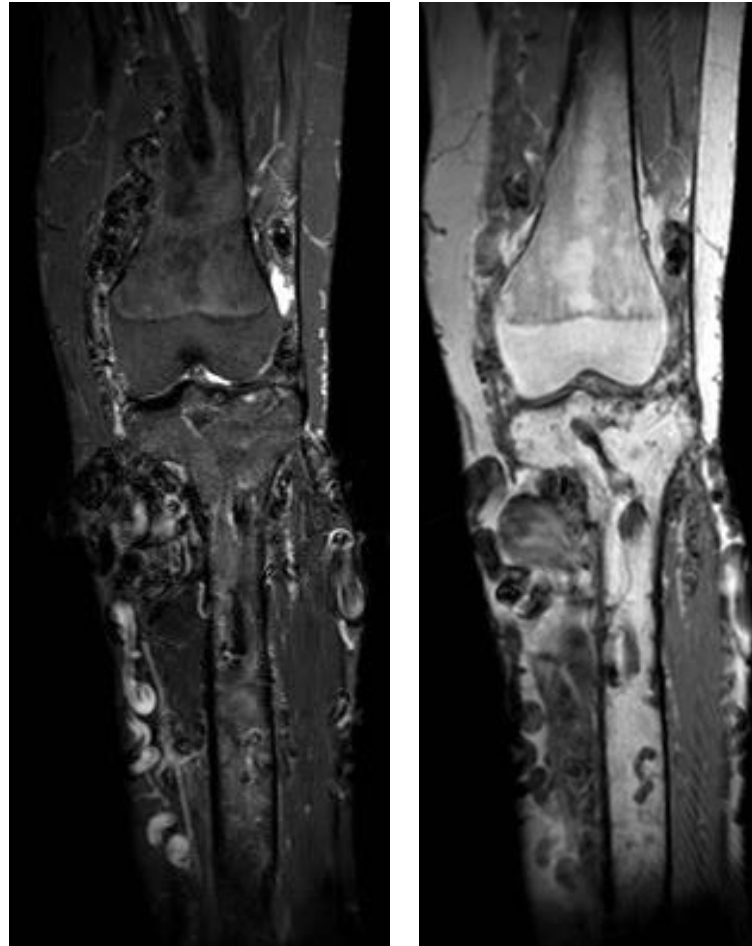
### – Evaluate flow imbalance

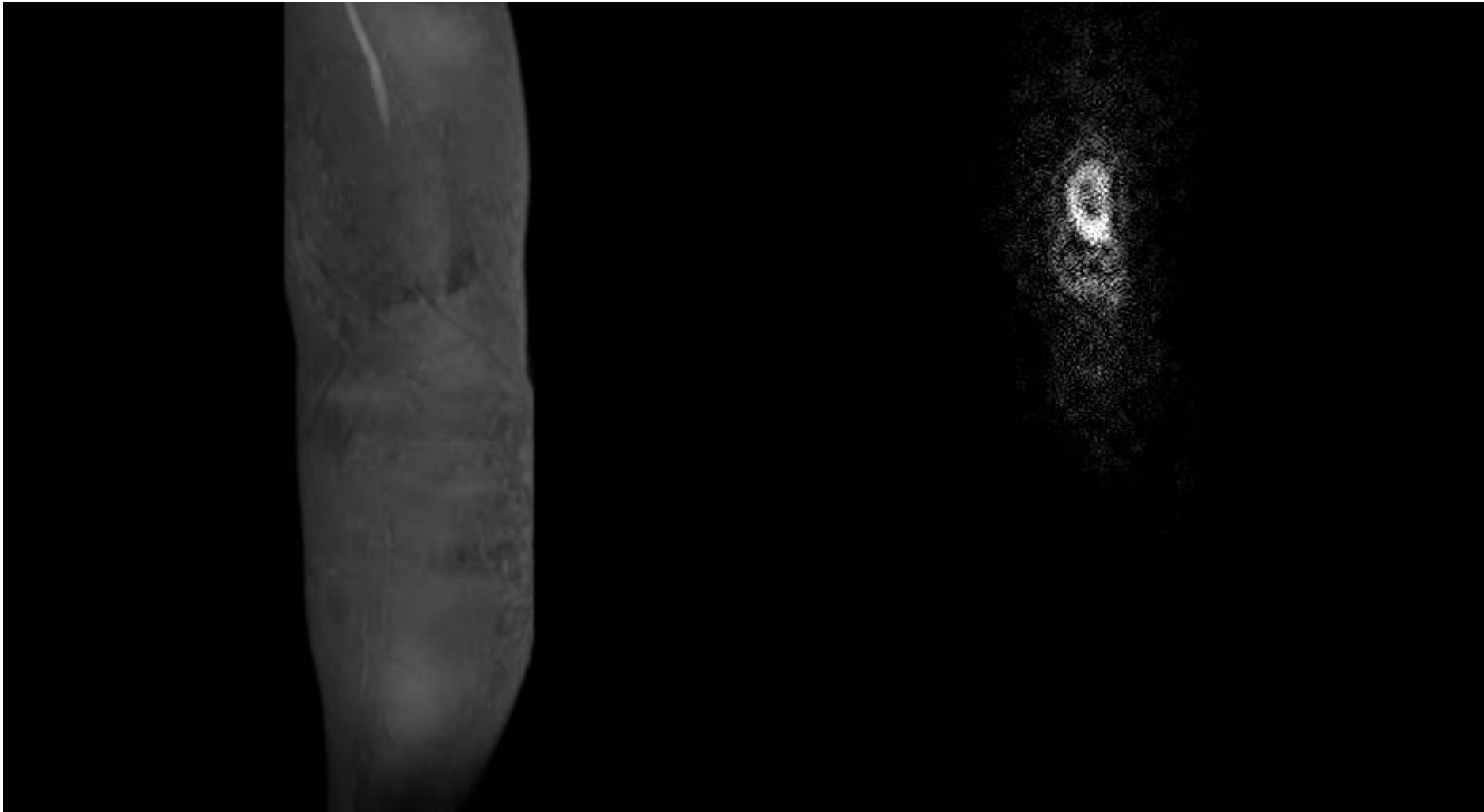
- Arterial feeder
- Venous drainage
- Normal arteries distal to AVM +++



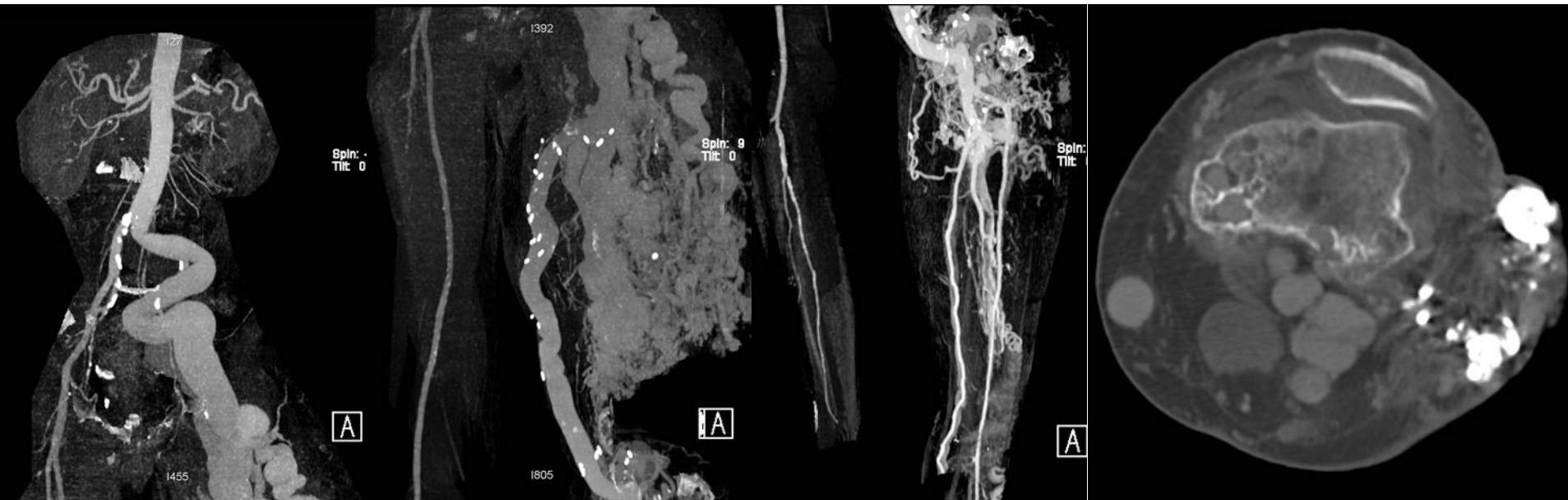
# MRI

- Flow void
- T1-T2
- 4D angio
- High resolution
- steady state





# CT-angiography



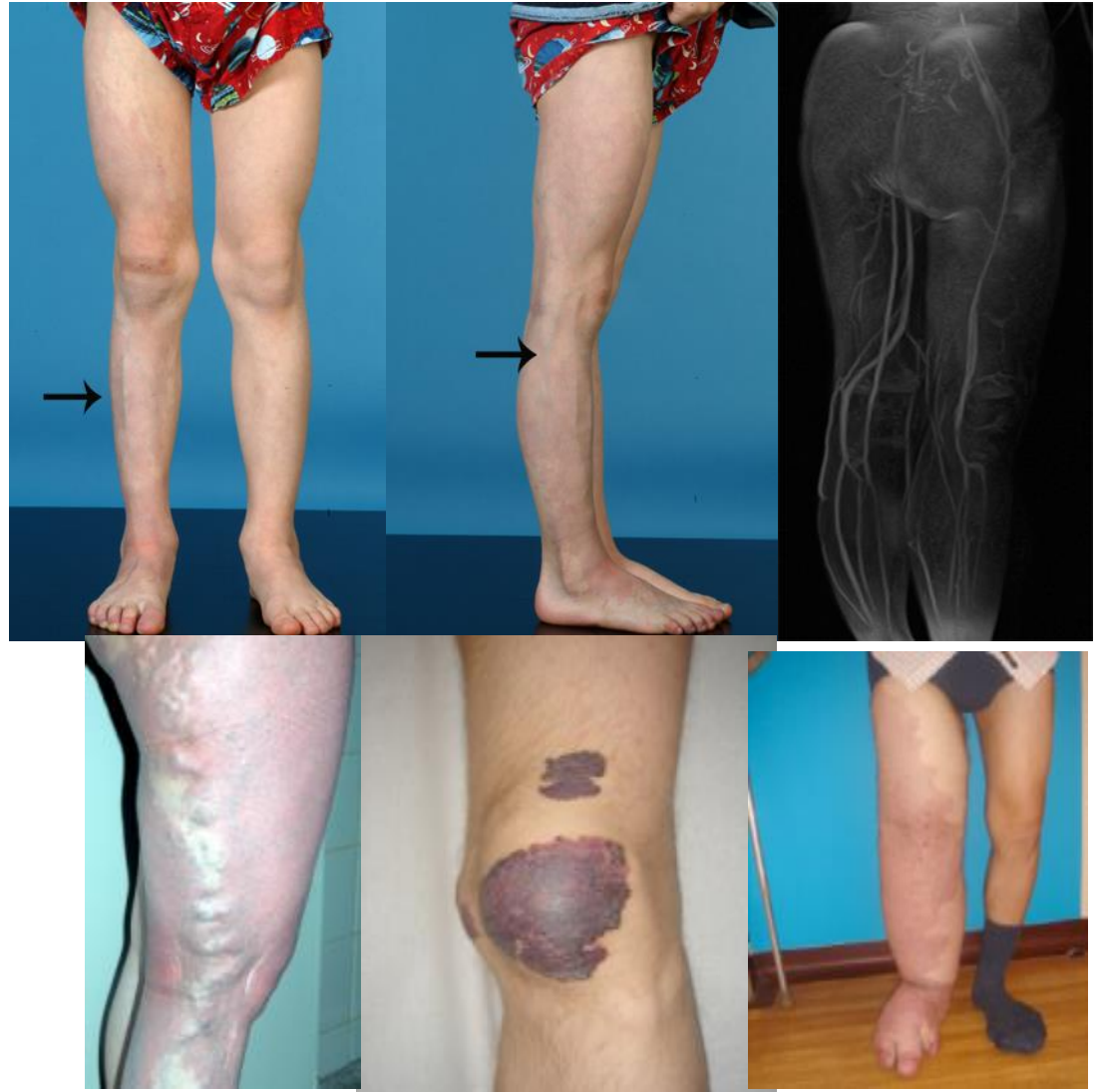
Arterial aneurysm and bone destruction

## Vascular malformations associated with other anomalies

|   |  |                   |
|---|--|-------------------|
| Klippel-Trenaunay syndrome:                           | CM + VM +/- LM + limb overgrowth   |                   |
| Parkes Weber syndrome:                                | CM + AVF + limb overgrowth   | <a href="#">G</a> |
| Servelle-Martorell syndrome:                          | limb VM + bone undergrowth   |                   |
| Sturge-Weber syndrome:                                | facial + leptomeningeal CM + eye anomalies<br>+/- bone and/or soft tissue overgrowth | <a href="#">G</a> |
| Limb CM + congenital non-progressive limb hypertrophy |  |                   |
| Maffucci syndrome:                                    | VM +/- spindle-cell hemangioma + enchondroma   |                   |
| Macrocephaly - CM (M-CM / MCAP)                       |  | <a href="#">G</a> |
| Microcephaly - CM (MICCAP)                            |  | <a href="#">G</a> |
| CLOVES syndrome:                                      | LM + VM + CM +/- AVM + lipomatous overgrowth   | <a href="#">G</a> |
| Proteus syndrome:                                     | CM, VM and/or LM + asymmetrical somatic overgrowth                                   | <a href="#">G</a> |
| Bannayan-Riley-Ruvalcaba sd:                          | AVM + VM + macrocephaly, lipomatous overgrowth                                       | <a href="#">G</a> |

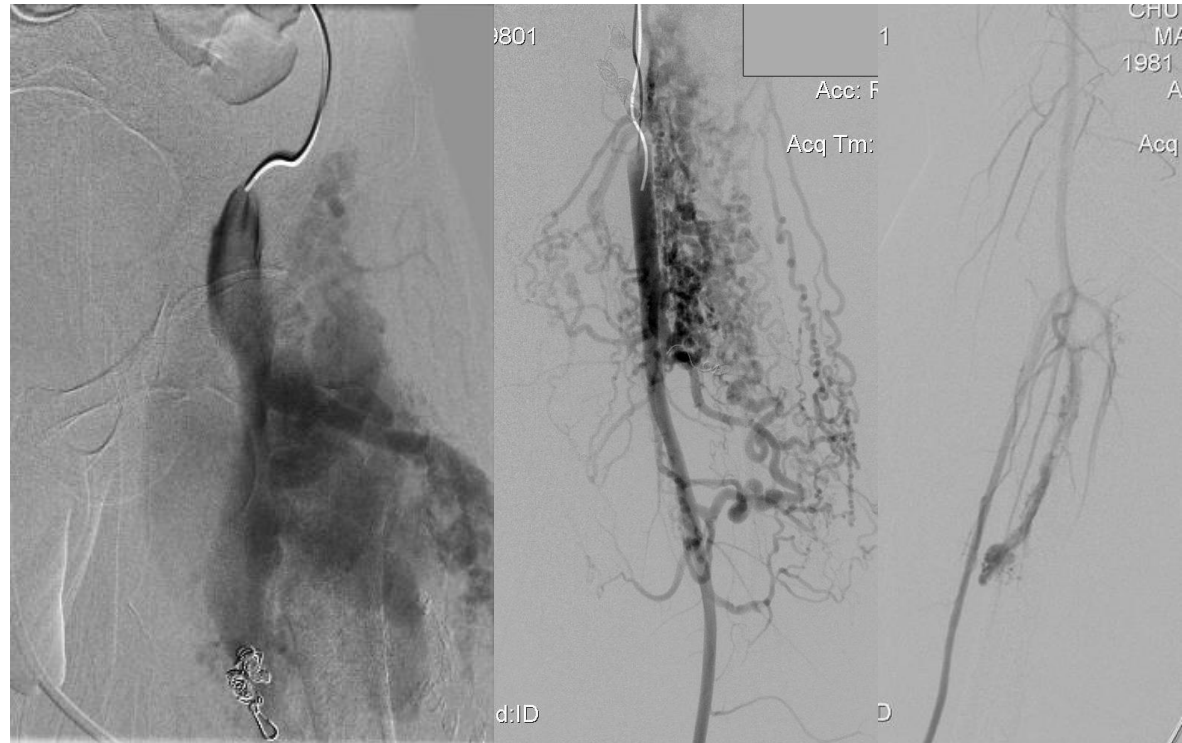
# Klippel trenauay

- Limb hypertrophy
- Cutaneous angioma
- Venous and or lymphatic
- R/O hypoplasia deep venous system
- Sclerosis of varicose vein



# Parkes Weber

- AVM
- Cutaneous angioma
- Limb hypertrophy



# Conclusion

- Make the diagnosis
- Clinical examination, Doppler, MRI
- Give a prognosis that will influence therapeutic planning
- Multidisciplinary approach
- IR pivotal role
  - Must be involved in clinical evaluation, imaging work-up and follow-up